Q Bullous:

1- Skin as an immunological organ.
2- Dermoepidermal junction.
3- Structure of desmosome.
4- Basic principles of desmogleine compensation theory.
5- Site of bullae in : bullous pemphigoid, pemphigus, dermatitis herpetiformis, subcorneal pustular dermatosis.
6- Pathogenesis of cutaneous lesions in mucocutaneous Pemphigus vulgaris
7- Paraneoplastic pemphigus: etiopathogenesis, CP, diagnosis, ttt
8- IgA pemphigus.
9- Treatment of a case of pemphigus.
10- Cicatricial pemphigoid.
11- Dermatitis herpetiformis: how to confirm diagnosis, associated disorders, therapeutic options.
12- Specify the types of epidermolysis bullosa & discuss epidermolysis bullosa dystrophica.
13- Mention the major diagnostic clinical criteria of epiermolysis bullosa acquisita
14- Nail changes in epidermolysis bullosa dystrophica
15- Mention the defect in junctional epidermolysis bullosa .
16- Epidermolysis bullosa pruriginosa
17- Hailey Hailey disease.
18- Describe the immunoflorescence findings (direct & indirect) in autoimmune vesiculobullous diseases.
19- Discuss clinical features of bullous diseases are not enough for diagnosis.
20- Cutaneous manifestations of antiphospholipid syndrome.
21- Chronic bullous disease of childhood.
22- Compare Pemphigus vulgaris versus Paraneoplastic pemphigus .
23- Compare between bullous pemphigoid & epidermolysis bullosa acquisita.
24- Compare between : p. vulgaris, p. vegetans, p. foliaceus & p. erythematosus: clinically & histopathologically.
25- Compare between Hailey Hailey disease & pemphigus vegetans.
26- DD of bullous diseases.
27- Scarring bullous eruptions: etiology & diagnosis.
28- Histopathological DD of subepidermal bullae.
29- Bullous eruption of neonate.

سؤال رقم ①

* Skin as an immunological organ?

- Skin act as a barrier that protect the body from entrance of pathogens.

- Continous turnover of the horny layer of the skin.

- Keratenocytes of the epiderms has an immunological functions as they secrets inhibitory cytokines

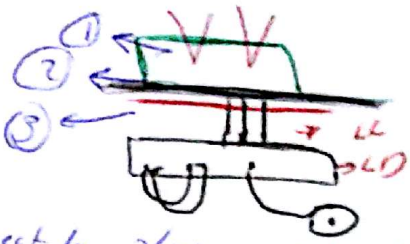
- Langerhanse of the epidermal cells. They act as antigen presenting cells that attack the foreign antigens & travel through the lymphatics to presents them to T lymphocytes to start there action.

- Keratenocytes secrets antimicrobial peptieds that has antimicrobial effect: as defensins & ~~Cathelethedins~~ Cathelicidins.

Derma-epidermal Junction DEJ
Basement membrane Zone BMZ

Function:-
1. Adhesion of Dermis and epidermis
2. Mechanical support
3. Selective permeability, physical barrier
4. Mesenchymal-epidermal interaction

Composition By electron microscope



[1] Basal cell plasma membrane → hemidesmosome (HD)
① inner plaque (connect to IF) ② outer plaque (connect to plasma membrane)
③ Sub basal dense plate

[2] Lamina lucida → lucent area contain Anchoring filaments (HD → LD)

[3] Lamina densa → electron dense band

[4] Sub lamina densa → Fibrillar zone contain Anchoring
- fibrils (Not filaments) which may loop back to LD or inserted to electron dense plaque (Anchoring plaque)

= Molecular Components =

[1] Hemidesmosome →
→ Inner plaque → plectin 500 (KD)
BPAG1 230 (KD)
→ Transmembrane components → BPAG2 180 (KD)
→ α6β4 Integrin

[2] Lamina lucida → BPAG2 (extracellular segment)
- β4 extracellular domain of α6β4 Integrin
- Anchoring filaments contain IgA (linear) + Laminin (5)
- Laminin 5, 6 connected together By disulfide bond
- Uncein - Fibronectin.

[3] Lamina densa:-

① Collagen IV → Non Fibrillar Collagen  red shaped with 4 domains, it works as mesh-like support of LD

② Nidogen → Small, sulfated glycoprotein 150 kd نيدوجين

③ Heparin sulfate proteoglycan → glycoaminoglycan. regulate migration of charged cells throughout BM

④ Chondroitin-6-sulfate → the same as HSP

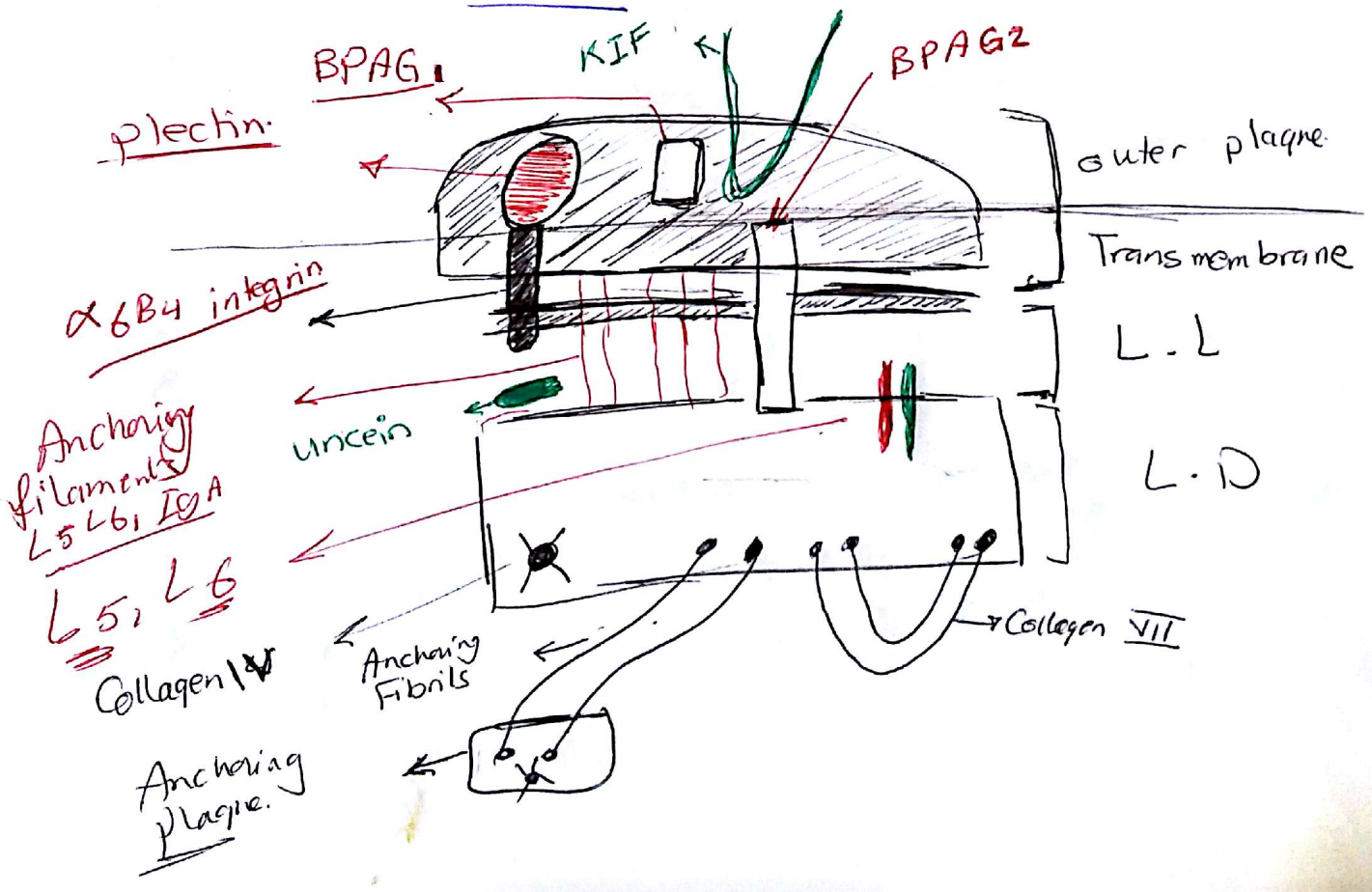
⑤ Laminin 5, 6

[4] Sub lamina densa:-

① Anchoring fibrils → Collagen VII → The target Antigen in BSLE.

② Interstitial Collagen → Collagen I, III → Papillary dermis

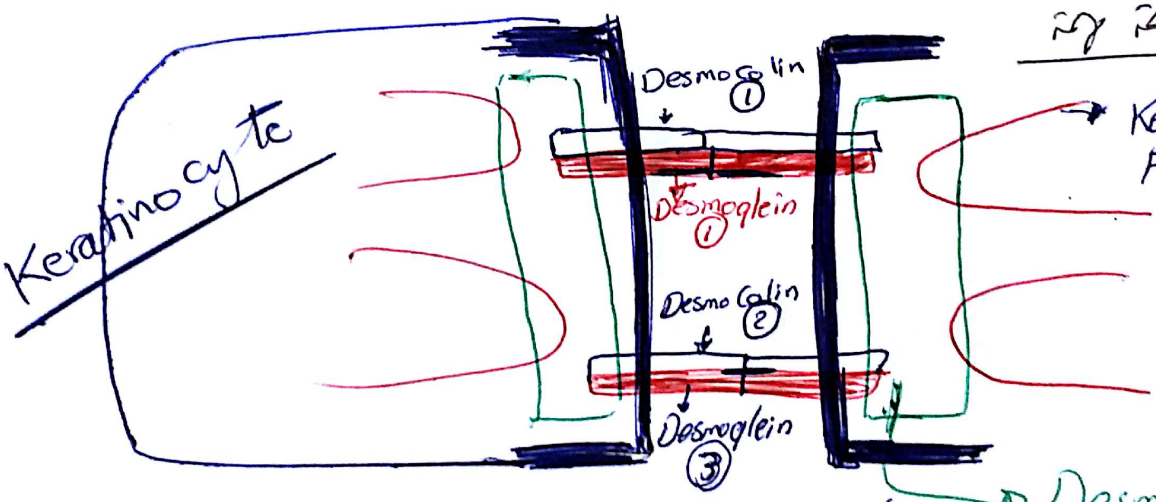
③ Microfibrils → Fibrins + elastins



- Desmosome -

- highly electron dense material Contain of proteins.
- Connect Keratinocytes with each other (3) groups of protein
  1. Intermediate Filaments → Keratin 5, 14
  2. Plaque Contain of plakins Desmoplakin I, II
  3. Core Cadherins → desmoglein 1, 3 & Desmocolin 1, 2
     (Transmembran proteins)

NB Cadherin means Ca^+ dependent adhesion molecules



NB Desmoglein 1 in upper epidermis
Desmoglein 3 in lower epidermis.

Question [4]

Desmoglein Protein

(Desmosome + glu)

Type I — Type III

Desmoglien 1 & Desmoglin 3

Can Compansate each other if there is Deficiency in either.

But the other must Present in large amount to Compansate (Sufficient).

Normally

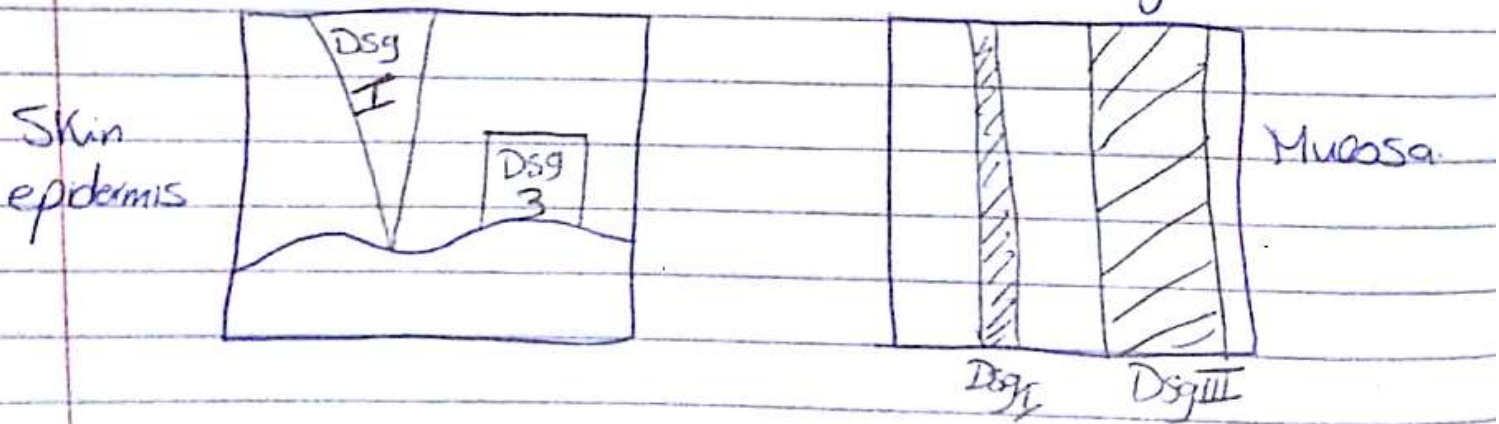
Desmoglein I present ① in epderms of Skin but more in Superficial layer

② through all mucosa but in lower level than Dsg3.

Desmoglein 3 present in

① lower part of epdermis (basal & suprabasal)

② Through all mucosa in large amount.



So if Dsg3 ↓↓ in epidermis ⇒ Dsg1 Compansate it.

Application on Pemphigus Vulgaris

1) Mucosal Dominent pemphigus Vulgaris :-

Dsg 3 only is Affected

So → in Skin Dsg 1 Compansate the Defeciency.
↳ in Mucosa Dsg 1 Can't Compansate the Defeciency as it present in Small amount.

∴ ⇒ Skin will be intact.
⇒ Mucosa will be affected. → (Mucosal erosion).

2) Muco-Cutaneous Subtype of P.V

Dsg 1 & Dsg 3 are affected.

So No Compansation
∴ Both Skin & Mucosa affected.
⇒ Mucosal erosion & Skin blisters.

⑤ Site of bulla in:-

Ⓐ Bullous pemphigoid

* In non-bullous phase: there is no blister just eosinophilic spongiosis and dermal infiltrates of eosinophils (at Flexual aspects of the Limbs and lower trunk)
* In bullous phase: Subepidermal blister with eosinophils at the same site

Ⓑ pemphigus:-

* pemphigus vulgaris: intra epidermal blister just above basal cell layer (supra-basilar)
* pemphigus vegetans: Supra-basilar blister

Ⓒ Dermatitis herpitiformis

Sub-epidermal clefts with collection of neutrophils within dermal papillae

(2)

(D) Sub corneal pustular dermatosis

pustules are located sub-corneally in the upper epidermis in IgA pemphigus

⑥ pathogenesis of Cutaneous lesions in mucocutaneous pemphigus vulgaris:-

in muco-cutaneous type the target protein in the desmosome is desmoglein 3 and desmoglein 1 ξ

* Desmoglein 1 :- is expressed in superficial layers of the epidermis of the skin.

* Desmoglein 3 :- is expressed in the lower part of the epidermis of the skin.

* while Desmoglein 1 and 3 are expressed together in the mucosa but Dsg 1 have a lower level than Dsg 3

* if one desmoglein is present enough, it can compensate for the loss of the other and because of their

there is no skin lesions in mucosal-dominant type but in muco cutaneous type there is affection of both Dsg1 and Dsg 3 leading to affection of both skin and mucosa

Q7 Paraneoplastic Pemphigus

التاريخ :

Def.

It is a type of pemphigus associated with underlying (Neoplasms)

Pathogenesis

(1) underlying neoplasms → Benign / malignant

(2) - Most Common neoplasms

2/3 of pt- - Non-Hodgkin lymphoma
- Ch. lymphocytic leukemia
- Castleman's disease
- Thymomas (Benign or malignant)
- Sarcomas

(3) Target protiens

polymorphic many antigen [in desmosome & BM

* Desmoglein 3, 1
* Desmoplakin I, II
* Envoplakin
* periplakin
* BP AG1
* plectin
* A2ML1

التاريخ :

Clinical picture

(1) Stomatitis (2) mucosal
(3) cutaneous (4) systemic

(1) Stomatitis

- erosion & ulceration
- extremely resistant to ttt
- most main symptoms of PNP
- earliest presenting sign
- after ttt it persists

(2) mucosal affection

- pseudo-membranous (conjunctivitis → scarring → obliteration of fornices
- oesophageal - vagina - penis - labial may ◦ Ca

(3) Cutaneous

- polymorphic appearance
  - (a) erythematous macule
  - (b) tense blister
  - (c) erythema multiforme
  - (d) lichenoid eruption

(4) Systemic

bronchiolitis obliterans
↓
fatal
↓
respiratory failure

التاريخ :

Histopathology

Cutaneous lesion → Polymorphism

1- pemphigus vulgaris like → supra basilar acantholysis

2- E.M like → keratinocyte necrosis with lymphocytes within the epidermis

3- lichen planus like → basal cell liquefactive degeneration

Immunofluorescence

DIF → intracellular and subepidermal IgG

Investigation to detect underlying neoplasm

1- CT chest, abdomen, pelvis
2- CBC
3- Flow cytometry
4- lactate dehydrogenase (LDH)
5- serum protein

D.D

[1] cutaneous lesion	[2] mucosal lesion
* Pemphigus vulgaris * Stevens - Johnson S * Erythema multiforme * Cicatricial pemphigoid * Lichen planus * Graft versus - host D (GVHD)	* Resistent HSV infecti * other viral infection * Stomatitis due to chemotherapy

Prognosis

Poor

Treatment

[1] Benign tumor → surgical excision

[2] malignant tumor → Chemotherapy.

Q 8 IgA Pemphigus

التاريخ :

Def

it is a type of pemphigus characterized by :-

1- IgA autoantibodies « immunopathology »

2- neutrophils « histologicaly »

3- Response to dapson « therapeutically »

Subtypes

according to level of intra epidermal pustule

1- subcorneal pustular dermatosis «SCPD» like type

pustule → subcorneal → in upper epidermis

2- Intra epidermal neutrophilic «IEN» type

suprabasilar pustule → lower or entire epiderm

Pathogenesis

1- SCPD → Desmocollin b

2- IEN → Unknown antigen

التاريخ :

Age

middle age or elderly.

Clinical picture

★ Symptoms

1- pruritis is the main symptom

★ Signs.

1- flaccid vesicle or pustule
2- on erythematous or normal skin
3- pustule coalesce to form annular or circinate pattern
↓
((Sunflower- like configuration of pustule))
4- Common site axilla, groin but Trunk & extremities may be

Notes

1- general condition is ((Good))

2- mucous membran involvment ((Rare))

التاريخ :

Histopathology

1- SCPD → pustule is «Subcorneal» in upper epidermis

2- IEN → «Supra-basilar» pustule in lower or entire epidermis

Immunofluorescence

1- Direct IF → IgA «deposition» on cell surfaces of epidermal keratinocytes

2- Indirect IF → «circulating» IgA autoantibodies

D D

S + D ↓ → 3P + 1D + 1L

1- pemphigus foliaceus
2- bullous impetigo — 3P
3- pustular psoriasis

4- dermatitis herpetiformis — D

5- linear IgA bullous dermatosis — L

6- subcorneal pustular dermatosis — S

التاريخ :

Prognosis

chronic indolent course

Treatment

1- Dapson is the drug of choice (علاج الاختيار)

2- response 24-48 hours.

3- if dapson failed

- sulfapyridine
- acitretin

4- if ↑ failed

- low-medium dose prednisone
- photochemotherapy (PUVA)

or - colchicine

.. سؤال رقم ٩ ..

tt of a case of pemphigus :.

Choice of tt: depends on:

① severity → mild → Corticosteroids
→ moderate/ sever → Cs + immunosupressants.

② patient (some patients Corticosteroid are contraindicated as elderly, sever Hypertension)

③ Drug (the cost as an example)

First line therapy:

* Prednisolone
  - 1mg/kg/day.
  - Add (H_2 blockers, vit D, calcium).

2nd line (If no response to Cs (or) Contraindication of Cs))

* MyCophenolate:
* - 2-3 gm/day
  - Continous CBC
* Azathioprine
  2-4 mg/kg/day

3rd line (If no respons for imm... (or) Contraindication for imm...)

* Rituximab (anti CD20)
  400mg/kg/day for 5 days

* Intravenous immunoglubulins:
  375 mg/m² once/week for 4 weeks

* plasmapharesis:
  2 times / week.

* in sever cases (pulse intravenous steroid therapy)
IV (Methy) prednisolone 1gm/day over a period of 2-3 hr for 5 day. then shift to oral ttt. If no response repeat the pulse therapy once.

monitoring: → clinically: ⇒ No of new blisters
⇒ rate of healing.
→ laboratory: → IF & (EIISA)
↘ to adjust the dose of Cs if there is response

Tapering of the dose: (If response)

- If Cs alone Taper the dose as (25% of dose reduced every 2 weeks). ~~5-10 mg/week~~ 5-10 mg. every 2 weeks.

- If Cs + Immunosupressant.
  - Taper the dose of corticosteroid as before.
  - ~~If the dose of Cs~~
  - If the dose of 5-10 mg/day reched → Taper the Immunosupressant.
  - maintain the patient on 5mg/day of Cs or every successive day.

10- Bullous - Cicatricial Pemphigoid

def rare autoimmune dis involve both (mucous mem / skin) esp (oral & conjuctival m.m) & result in scarring.

incidence middle / old age. **age** 50-80 y

cause chronic course

types

	I (anti laminin) B	▷ II ocular. P.	III	IV
Target Ag.	⊙ laminin 322 ⊙ laminin 311	- B4 subunit of ($\alpha 6 B4$) integrin	BP 180	. unclear
Involvement	. associated ↑ NGII Tumor incidence, lung endometrium. . Tumor develop after 2 years of skin lesion	. Pure or Predominant Ocular dis	. affected skin & m.m.	. m.m involvement c/out skin involvement

clinical symptoms

- ▷ **Ocular sym**
  - . conjuctivitis → entropion, fusion (Palpebral / bulbar) conj.
  - . trichiasis → corneal neovascularization.
  - Scarring.
  - . sym blepharon
  - . blindness

- **skin lesion (25+30%)**
  - . mostly involved → scalp, face, neck upper Trunk.
  - . lesion → erythematous plaque → recurrent blister formation (eros
  - . No → limited
  - . scalp CB → scarring alopecia.

Histology

IIF → 20% → binding to roof (epidermal side).
↳ type I → binding to dermal side.

DD
- linear IgA dis.
- subepidermal bullous dis.
- ant P 200 pemphigoids.
- Bullous LE. - EBA.

B10

III I local → (VIP)
↳ skin lesion → Topical steroid.
oral lesions → Topical steroid < mouth wash. gel. orabase base
↳ AB mouth wash.
↳ good hygiene.
nasal, Pharyngeal, oesphageal → steroid sprayer, inhaler.

II Systemic III
1st line - Dapsone (50-150mg/d)
2nd line - Cyclophosphamide (1-2 mg/kg/d)
↳ DOC for Progressive severe autoimmune dis.
↳ ± steroids.
3rd lines → azathioprine (2mg/kg/d)
(or) ↳ Mycophenolate (2g/d).
(or) ↳ steroids (not alone).
↳ not so effective.

Others → sulfapyridine, tacrolimus, cyclosporine
↳ AB → tetracycline.

III Surgical III follow medical therapy control of dis.
for severe scar of eye, larynx, oesophagus

11 - Bullous- Dermatitis herpitiform.

Def chronic autoimmune Bullous dis.

epi - age → all ages (children c/p as adult c/p)

genetic → ↑ incidence of HLA DQ 2.
HLA B8.

Cause → chronic

diagnosis → @ Sym → severe Pruritis, symmetrical, polymorphic.
erythematous Plaques, vesicles, urticarial wheals
site → on extensor surface of limbs, shoulders, buttocks
± Palm involvement
healing → scar, Pigmentation
oral lesions → No
b) history → ingestion of gluten or iodine diet.

Investigation

HP → biopsy → best Perilesional skin
↳ • subepidermal vesicle
• Papillary neutrophilic microabscesses in dermal Papillae.
• dermal Perivascular neutrophilic infiltrate & nuclear dust.

EM → lamina lucida skin cleavage.

DIF → biopsy → Perilesional skin.
↳ granular IgA (IgA1 class) deposit of dermal. P.

serum finding → IIF → circulating anti BMZ Ab → -ve.
↳ auto Ag → epidermal transglutaminase.
↳ Tissue transglutaminase
↳ Ab → anti reticulin.
anti Thyroid Ab.
anti gliadin Ab.
IgA immune complexes

Pathogenesis

13/11

defective mucosal barrier → passage of gluten macromolecules → lamina propria → ⊕ immune response → IgA I Ab → immune complexes release in circulation.

→ deposited in dermal papillae of the skin → release of chemo attractants → ⊕ neutrophil attraction ⊕ to upper dermis → release of Pharmacologic subs + Protease → L.L separation & vesicle formation

Tt 1st line Dapsone (100-200 mg/d).
- Therapeutic test → significant response in 1st 48h.

2nd line → sulfapyridine (1-2 mg/d).

→ modify life style → gluten free diet → benefit → ↓ skin lesions, less dapsone twice. → Protect Pt from lymphoma.

DD IgA bullous dermatosis.

Hailey ⓐ (17) - Hailey disease (fBP).

AD - +ve family history - age (15-25 yr)

C/P: * Small flaccid vesicles on erythema base

* on sides, nape of neck, axilla, groin
circinate pattern, pruritus - Healing ē hyperpigm
- rare mm affection

*** Exacerbating factors**: Heat, Trauma, bacteria, candida inf

*** Pathogenesis**: AD - mutations in ATP2C1 gene
→ interfering ē Intracellular Ca++ signaling.

Comp.: * Infection → bacterial - viral - fungal
* malignant transformation → SCC.

Clinical subtypes: ① Segmental Type 1: mosaic distr of disease

② " Type 2: Granulomatous seg area of more pronounced involvement.

H-P: * Suprabasal lacuna → vesicles - bulla
* villi protruded in the bulla.
* acantholysis more extensive but less severe than P.V - few dyskeratotic cells.

DIF, IIF → -ve cas it is genodermatosis (~~bullous~~ bullous) not Immunobullous.

TTT - dapsone: 50-150 mg daily - TTT of choice
sulphapyridine (alternative).
* BBUVB, NBUVB, PUVA, RePUVA

N.B Steroids ineffective even in large doses

Q. - Describe the immunofluorescence finding (DIF - IIF) in Autoimmune Vesiculobullous disease

	DIF	IIF
* P. vulgaris * P. vegetans * P. foliaceus * P. erythematosus * Paraneoplastic Pemphigus (PNP)	- inter cellular IgG or C_3	circulating IgG Aut Antibodies against desmosomal cadherins
* IgA Pemphigus	* IEN type: IgA deposit in lower epidermal cell surface * spD type: IgA deposit in upper epidermal cell surface	circulating IgA Autoantibodies to epithelial cell surface.
* Bullous Pemphigoid	liner IgG C_3 along DEj	circulating AutoAntbody IgG against DEj SSS (roof)
* Cicatricial Pemphigoid	liner IgG, C_3 along DEj	as Bullous Pemphigoid.
* Herpes Gestationis	as Bullous Pemphigoid	as Bullous Pemphigoid
* epidermolysis bullosa acquisita	as B. P (Bullous Pemphigoid)	- IgG against BMZ SSS (floor)
* Dermatitis herpitiform	- granular IgA deposit at dermal Papilla	- -ve (negative)

18-2

	DIF	IIF
* linear IgA bullous dermatosis (LABD)	- liner IgA ± C_3 at BMZ	- circulating Autoantibody IgA SSS (roof)
* Ch bullous dermatosis of childhood (CBDC)	- liner IgA along DEJ	- circulating Autoantibody rgA.

2a - Cutaneous Manifestation of Antiphospholipid syndrom.

1- livedoreticularis
2- Acrocynosis
3- ulceration
4- Necrosis
5- Raynaud's phenomenon
6- Capillaritis
7- Purpuric - cyanotic Macules
8- Nodules
9- digital ischema - gangrene
10- Blue toe
11- Thrombo phlebitis
12- Hemorrhage
13- Porcelin white scar - atrophic blanch
14- splinter Hemorrage.

21 - ch bullous dermatosis of childhood (CBDC)

* Age → 3-5y . It last for few years ē spontaneous resolution befor Puberty
* clincaly → pruritic vesicles and bulla on erithematous or normal skin affect genitalia - buttock - thigh - perioral area.
oral lesion occure in 50% of Patient.
Bulla arranged in annular Pattern (cluster of jewels)
healing occure ē hyperpigmentation
* histoPathology → subepidermal bulla ē predominance of neutrophil or with esinophil.
* DIF → liner IgA along DEJ
* IIF → circulating IgA Ab BMZ antbody
* Antigen → separation in LL → Antigen BPAG2
" " SLd → " type 7 collagen
* TR → - sulphapyridin 250mg - 3gm daily
- dapson 1-2 mg/kg.

P. vegetans
Paraneoplastic pemphegus.

Age — pediatric / adult

pathogenesis — Ab:- IgG auto anti bodies : C from plakin containing tail
Ag:- D3, D1, desmoplakin I, II, envoplakin, periplakin, BPAg 1

C/P — mucosa: severe mucositis & Erosion, Crustation, Clip, lat aspect of Lingual
skin + polymorphus skin lesion
- PV like - EM like - Bp like - lichen like
EM like

Histo pathology
- supra basal acantholysis.
- keratinocyte necrosis.
- vacular interface change & sparse perivascular infiltrate
- band like infilteration

Immuno pathogy

DIF — IgG & Complement deposits in ICs & not granular linear complement deposition along BMZ

IIF — Circulating IgG auto antibodies bind to epithelial cell surface (as in pemphegus & to rat bladder (simple, Col, transitional epi)

tumors
1- Non Hodjkins lymphoma
2- ch. lymphocytic leukemia
3- Castle man's disease (1st in children 3rd in adult)

Date:
Subject:
No.:

pemphigus vulgaris

Age :- 40 years old middle east 50-60 urope

- pathogenesis :- Ab IgG (auto antibodies)
Ag D_1 (skin), D_3 (mm).

- C/p :- Skin- Bulle (large, flaccid, containing clear, turbid, hge sero purulent)
- rupture to leave denuded areas
- heal with hyper pigmentation, no scaring
- +ve Nikolioki sign +ve Asboe-Hansen sign

mm :- painful erosions on soft palate, buccal mucosa
- nasal mucosa, oropharyngeal, oesophageal.

- Histopathology :- - supra basal acantholysis - esinophils inside bule
- raw of tomb stone
- dermal papille projecting upward into bulle
- no keratinocyte necrosis.
- if early esinophilic spongiosis

- immunopathology

DIF :- - of perilesional skin → intercellular IgG & or C_3 along keratinocyte mems in 100% of patient

IIF - Circulating IgG auto antibodies (IgG1, IgG4)
- titre is related to disease activity

Association :- - MG
- thymoma.

Subtype P. vegetans :- reaction of skin to auto immune insult → Blister (Neumann type)
- pustule (in hallopeau type) → hyper trophic vegetating lesions mainly affecting flexures
- if tongue affected → cicitriform tongue (prenolata sign)

HP - marked acanthosis & papillomatosis - intra epidermal esinophilic micro abscess

q 22_2

Date:
Subject:
No.:

Bullous Pemphegoid.

Age:- > 60 year ... no sex predilection

pathogenesis: Ab : IgG auto antibodies

Ag :- BP Ag 1 . mainly - BP Ag 2 (c)

C/P: skin large tense blisters on normal or erythematous urticated plaques skin, mainly on flexures but may be generalized when resolving → hyperpigmentation & not scarring may heal spontaneously with new lesions appear - ve Nikolsky & Asbo-Hansen sign. no HPS lesion

mm mm : oral lesions in upto 90%. /no

Histopathology: sub epidermal bulla with eosinophilia within blister

Immuno DIF: IgG & or C_3 along DEJ (within LL).

IIF: circulating IgG auto antibodies against BP Ag
The titre not related to disease activity

Salt split skin technique: auto antibodies will react with the epidermal side

Date:
Subject:
No.:

Epidermolysis bullosa acquisita

Age adult 40–50 yr. male or & out Family history

pathogenes Ag :- type VII Collagen (anchoring fibrils)

Ab :- IgG auto antibodies

C/P trauma - induced non-inflammatory tense

(generalized variant) bulla → heal with scarring & milia

- in trauma - prone extensor surfaces - elbow, legs back of hands, nail dystrophy

- mm. affection common

H/P

sub epidermal bulla with neutrophilia

upper dermis

Immunopathology

DIF linear IgG, C_3 at DEJ

IIF auto BMZ auto antibodies against type VII C

salt split skin test auto antibodies → dermal side of blister

Question 25

Pemphigus vegetans & hailey hailey Disease.

اللغز الشبه الجوهري انهم Both

1) Clinically → intertriginous area.
2) H/P → Acantholysis

والفرق بينهم بالـ → Direct immunofluorescence

	Pemphigus Vegetans	hailey-hailey Disease.
Cause	Auto-immune Disease. (inflammatory Type of P.V)	Autosomal Dominant genetic Disease
Pathogenesis	Circulating antibodies against Cell surface of Keratinocyte.	- Mutation ATP2CA gene on chromosome 3q21 encoding [SPCA1] in golgi body [human - Secretory Pathway Ca^{+2}/Mn^{+2} ATPase]
Clinically	- Erosions with vegetations or Papillomatous Proliferation	- Painful erosion. - Pruritis - Malodor
Site	- Intertriginous Areas (flexures) - Scalp & face are common.	- Intertriginous site - Scalp is less frequently
H/P	- Suprabasilar Acantholysis. - eosinophilic infiltrate	- Acantholysis without Dyskeratosis.
Direct IF	+ve Perilesional Skin. IgG ± C_3 on Keratinocyte cell surface.	- -ve.